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PHYSIOLOGIC DIAGNOSTIC TESTS IN CONGENITAL HEART DISEASE*

Definitive diagnosis of tetralogy of Fallot and related cardiac malformations is frequently possible by clinical methods alone. In complex cases, however, physiologic procedures are of help in the preoperative recognition of these malformations. These procedures consist of (1) catheterization of the heart, (2) measurement of the total pulmonary blood flow (pulmonary capillary flow), (3) a standard exercise test, and (4) oximetry. The main diagnostic aim of these tests is the recognition of three conditions which must exist if the Blalock operation, consisting of construction of an artificial ductus, is to be attempted.[1] These three conditions are (1) the presence of an interauricular or interventricular septal defect; (2) obstruction to flow in the pulmonary artery resulting in a mean blood pressure in the pulmonary artery of less than that in the aorta and its branches; and (3) an oxygen saturation of peripheral arterial blood of less than 88 per cent. All three conditions are present in tetralogy of Fallot, tricuspid stenosis or atresia with septal defect, and single ventricle with normal or decreased pressure in the pulmonary artery. Condition 2 is missing in Eisenmenger's complex. Condition 1 is absent in isolated pulmonic stenosis.

Catheterization of the heart. The procedure described in a preceding number of this series, entails the introduction of a plastic catheter through an antecubital vein into the right side of the heart and the pulmonary artery[2]. In some instances the catheter may pass through a septal defect into the left side of the heart and possibly into one of the pulmonary veins or the aorta. The greatest amount of information is derived from this procedure if these principles are followed: 1. The catheter should be moved under roentgenoscopic visualization only. 2. Blood pressures should be recorded from all chambers of the heart and great vessels. 3. The oxygen content of blood in the heart, pulmonary artery and vena cava must be measured. 4. The volume of blood flow through the systemic circulation, the pulmonary artery and intracardiac shunt, and the effective pulmonary blood flow should be calculated. The effective pulmonary blood flow is the volume of mixed venous blood which, after having returned to the right auricle, is eventually aerated. If intubation of the pulmonary artery is impossible, calculation of the blood flow in the pulmonary artery, using the oxygen content of blood in the right ventricle, is of particular value in the recognition of pulmonic stenosis (Table 1). The analysis of the oxygen content and of volume flows is of help in recognizing and localizing septal defects. In the presence of a septal defect the volumes of blood flow in the pulmonary artery and systemic circulation are unequal. If the defect is interauricular, the oxygen content of right auricular blood is significantly higher than the oxygen content of blood from the superior vena cava. In a ventricular septal defect, the oxygen content of right ventricular blood is higher than that of right auricular blood.[3,4,5,6]

Determination of the total pulmonary blood flow (pulmonary capillary flow). This test is of aid in estimating the volume flow through collateral pulmonary vessels and in diagnosis of a persistent ductus arteriosus after the volume of flow in the pulmonary artery has been determined. The method involves the determination of the carbon dioxide content of incoming blood in the pulmonary capillaries and of blood in the pulmonary veins, and determination of the total production of carbon dioxide. The volume flow in the pulmonary capillaries may be estimated from these values by means of the Fick equation. The volume of blood coursing through the collateral vessels or through a patent ductus arteriosus may then be calculated by subtracting the volume of flow in the pulmonary artery from that in the pulmonary capillaries.

The standard exercise test. The main objective of this test is recognition of mechanical obstruction to blood flow in the pulmonary artery, (as in pulmonic stenosis) and of intracardiac shunts. During its performance (stepping up and down a step 20 cm. high, thirty times in one minute) the pulmonary circulation of a normal individual increases sufficiently to meet the respiratory demands of exercise. The normal individual shows, therefore, an increase in the ratio of oxygen consumed to the liters of ventilation during the test. In patients who have pulmonic stenosis, however, the obstruction limits the effective pulmonary blood flow and this ratio falls. In the absence of a septal defect, oxygen saturation of peripheral arterial blood remains constant during and immediately following the exercise. If a septal defect is present oxygen saturation of peripheral arterial blood declines.

Oximetry. The oximeter is an instrument which measures the oxygen saturation of blood in the peripheral arteries by means of a photo-electric cell attached to the pinna of the ear. Its main diagnostic value is in the continuous registration of the oxygen saturation during the standard exercise test. The oximeter is of particular physiologic interest in congenital heart disease since it also permits continuous recording of oxygen saturation of blood in peripheral arteries during inhalation of 100 per cent oxygen. In a normal individual, breathing pure oxygen results in full saturation of peripheral arterial blood within ten to fifteen seconds. In patients with intracardiac venous-arterial shunts, however, the saturation time is prolonged to from two to four minutes or longer, and full saturation is not reached[7]. The curves representing the increase in saturation vary in their slopes. This indicates that the speed of the oxygen uptake is dependent on the difference in oxygen gradient between blood in the pulmonary vein and the shunt. It is probable that the slope of these curves is a function of the initial oxygen saturation. The final height of the oxygen saturation is limited by the volume of blood in the venous-arterial shunt and its oxygen saturation on the one hand and by the volume of flow in the pulmonary capillaries and the oxygen saturation of blood in the pulmonary vein on the other[8].

Results. The results of these tests in various congenital heart lesions are illustrated in table 1. It may be seen that tetralogy of Fallot is characterized by a reduction of flow in the pulmonary artery and by an intracardiac shunt which is directed toward the

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left. The systolic pressure in the pulmonary artery is below that in the right ventricle which is above normal. In many cases of tetralogy of Fallot the flow in the pulmonary capillaries exceeds that in the pulmonary artery; this observation indicates an appreciable collateral circulation to the lungs. The oxygen saturation of peripheral arterial blood is determined not by the total pulmonary blood flow but by the percentage of mixed venous blood reaching the lungs. During the standard exercise test the oxygen consumed per liters of ventilation and the oxygen saturation of peripheral arterial blood decline. After the Blalock operation the effective pulmonary blood flow and the flow in the pulmonary capillaries increase, and the oxygen consumed per liters of ventilation usually rises during exercise.

In Eisenmenger's complex flow in the pulmonary artery is normal or slightly reduced. The intracardiac shunt may be directed from left to right or from right to left. The systolic pressures in the pulmonary artery and in the right ventricle are elevated. The diastolic pressure in the pulmonary artery is also above normal. These findings demonstrate increased resistance in the pulmonary vascular bed. During the performance of the standard exercise test the oxygen consumed per liters of ventilation rises but the oxygen saturation of blood in peripheral arteries decreases.

In patients with pure pulmonic stenosis the blood flows in the pulmonary artery and in the systemic circulation are equally reduced. The oxygen contents of arterial and ventricular blood are identical, since there is no intracardiac shunt. The systolic pressure in the right ventricle is higher than the systolic pressure in the pulmonary artery. During the standard exercise test the oxygen consumed per liters of ventilation falls, but the oxygen saturation in peripheral arterial blood remains constant.

Single ventricle with pulmonic stenosis is characterized by a large difference between the oxygen content of blood in the right auricle and the ventricle and by a reduction of the blood flow through the pulmonary artery.

In stenosis or atresia of the tricuspid valve, right auricular pressure is elevated. Since there is an interauricular septal defect, the oxygen content of right auricular blood significantly exceeds that of the superior vena cava despite the fact that the over-all shunt is from right to left. The intracardiac shunt is from right to left, and the oxygen consumed per liters of ventilation falls during the exercise test.

In the presence of an auricular septal defect the oxygen content of right auricular blood exceeds that of vena caval blood. With a ventricular septal defect the oxygen content of right ventricular blood exceeds that of right auricular blood. Right ventricular and pulmonary arterial pressures may be elevated. The intracardiac shunt may be directed toward the right or the left; when it is directed to the left, it probably indicates an increase in the pulmonary vascular resistance in certain cases of septal defect. During the standard exercise test the oxygen consumed per liters of ventilation increases and the oxygen saturation of peripheral arterial blood usually falls[9].

Normal blood pressures in the right ventricle and pulmonary artery are seen in a persistent ductus arteriosus. The oxygen content of blood in the pulmonary artery exceeds that in the right ventricle and flow in the pulmonary capillaries exceeds that in the pulmonary artery (Table 1).

Summary. Physiologic tests have been described which supplement the clinical procedures used in the diagnosis of congenital heart disease.

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Table 1

DIAGNOSTIC CRITERIA IN CONGENITAL HEART DISEASE*

	Catheterization and determination of pulmonary capillary flow			Exercise test and oximetry	
	Pressures, MM. of mercury	Analysis of gases in blood	Flows	Oxygen consumed per liter of ventilation	Oxygen saturation of peripheral arterial blood
Normal	RA = 5/0 RV = 25/0 PA = 25/8	RA = RV = PA within 0.5 vol. per cent	S = PA = PC =	Rises	Unchanged
Tetralogy of Fallot Preoperative	RV elevated PA lower than RV	O_2 in RV significantly higher than in RA	S exceeds PA	Falls	Falls
Postoperative	RV elevated PA lower than RV	O_2 in RV significantly higher than in RA	Effective and PC increased over Pre-Op.	Rises	Falls
Eisenmenger's complex	RV elevated PA systolic and diastolic elevated	O_2 in RV markedly higher than in RA	S may exceed PA flow or vice versa	Rises	Falls
Pulmonic stenosis	RV elevated PA lower than RV	RA = RV	S = PA; both reduced equally	Falls	Unchanged
Single ventricle with pulmonary stenosis	RA and RV elevated	O_2 in RV markedly higher than in RA (Greatest difference observed)	S exceeds PA	Falls	Falls
Tricuspid stenosis	RA elevated	O_2 in RA significantly higher than in SVC	S exceeds PA	Falls	Falls
Patent ductus arteriosus	RV = PA = normal	O_2 in PA significantly higher than in RV	PC exceeds PA	Rises	Unchanged
Isolated septal defects	Normal or elevated	RA significantly exceeds SVC, or RV exceeds RA	PA may exceed S or vice versa	Rises	Falls

*RA-right auricle; RV-right ventricle; PA-pulmonary artery; PC-pulmonary capillary; S-systemic circulation; SVC-superior vena cava.

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